# AN OVERVIEW OF RETROPERITONEAL TUMOURS

***Dissertation Submitted to***

THE TAMIL NADU DR. M.G.R. MEDICAL UNIVERSITY

***in partial fulfillment of the regulations***

***for the award of the degree of***

**M.S. BRANCH – I**
**GENERAL SURGERY**


**GOVT. STANLEY MEDICAL COLLEGE & HOSPITAL**
**THE TAMIL NADU DR. M.G.R. MEDICAL UNIVERSITY**
**CHENNAI, INDIA.**

**SEPTEMBER 2006**

# CERTIFICATE

This is to certify that the dissertation titled "**AN OVERVIEW OF RETROPERITONEAL TUMOURS"**of **Dr. K. RAJASEKARAN** in partial fulfilment of the requirements for **M.S. Branch – I (General Surgery)** Examination of the Tamilnadu Dr. M.G.R. Medical University to be held in September 2006. The period of study was from July 2003 to March 2006.

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# DECLARATION

I, **Dr. K. RAJASEKARAN** solemnly declare that dissertation titled, "**AN OVERVIEW OF RETROPERITONEAL TUMOURS"** is a bonafide work done by me at Govt. Stanley Medical College & Hospital during 2003-2006 under the guidance and supervision of my Unit Chief

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The dissertation is submitted to Tamilnadu Dr. M.G.R. Medical University, towards partial fulfillment of requirement for the award of **M.S. Degree (Branch – I) in General Surgery.**

Place : Chennai.

Date :

**(Dr. K. RAJASEKARAN)**

## ACKNOWLEGEMENT

I owe my thanks to the Dean, Govt. Stanley Medical College and Hospital, **Dr. M. VASANTHA, M.D.** for allowing me to avail the facilities needed for my dissertation work.

I am grateful to **Prof. Dr. D.R. GUNASEKARAN, M.S, FICS,** Professor and Head of the Department of Surgery, Govt. Stanley Medical College Hospital for permitting me to do the study and for his constant encouragement.

I am so thankful to our former unit chief **Prof. Dr. P.G. KOLANDAIVELU, M.S.** for his valuable guidance and suggestions.

I am extremely thankful to my unit Chief **Prof. V. SHRUTHIKAMAL, M.S.,** for her guidance and encouragement.

I owe my sincere thanks to our Assistant Professors **Dr. P. RAMANUJAM, M.S., Dr. G. MUTHUKUMARAN, M.S. and Dr. D. DURAI, M.S.,** for their valuable guidance and appropriate suggestions.

I thank my seniors colleagues, my fellow postgraduates and my junior colleagues, without whose help this study would not have been possible.

Last but not the least, my sincere thanks to all the patients who cooperated for this study, without whom this study could not have been possible.

# CONTENTS

# INTRODUCTION

Retroperitoneal tumours are uncommon heterogeneous group of tumours arising either primarily in retroperitoneum or representing metastasis from elsewhere.

The reported incidence of retro peritoneal tumour varies from 0.3 to 3%. Surgery remains the treatment of choice. But majority of the tumours are advanced at presentation and complete resection possible only in 40-70% of cases.

Our study at Stanley Medical College and Hospital, Chennai for the period of 2003 – 2006 deals with various presentation and features of retro peritoneal tumours. Both patient factors and surgical factors are taken into consideration.

# AIMS AND OBJECTIVES

**To study**

- **The age and sex incidence of retro peritoneal tumours**
- **The clinical presentation (Signs and Symptoms)**
- **The diagnostic modalities used in evaluation of retro peritoneal tumors**
- **The various types of treatment offered**
- **The immediate post operative complications**
- **The various histology of retro peritoneal tumours.**

# REVIEW OF LITERATURE OF RETROPERITONEAL TUMOUR
# ANATOMY OF RETROPERITONEUM

Retroperitoneum is an actual and potential space between the peritoneal cavity and posterior abdominal wall, containing structures of mesodermal, ectodermal origin with their embryonic remnants.

## *BOUNDARIES OF RETROPERITONEUM*

Retroperitoneum has superior, inferior, anterior and posterior boundaries.

Superiorly → $12^{th}$ rib and diaphragm

Inferiorly → Pelvic diaphragm and fascial of levator ani and
Coccygeus muscles.

Anteriorly → Posterior parietal peritoneum and the space
between the leaves of small and large bowel mesenteries.

Posteriorly → Vertebral column
Psoas muscles
Quadratus lumborum
Tendinous portion of transverse abdominus.

## *EMBRYOLOGY*

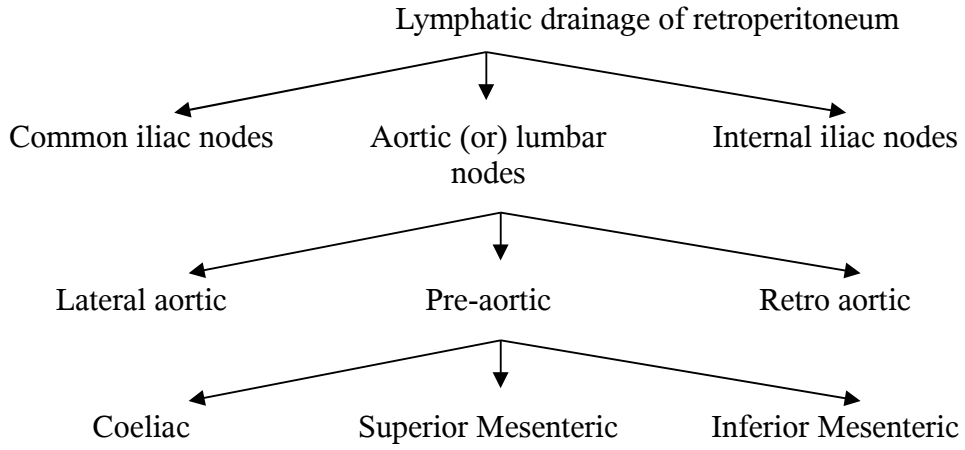
Retroperitoneum constitutes the tissues derived from the ectoderm, mesoderm and embryonal remnants.

## *CONTENTS OF RETROPERITONEUM*

Retroperitoneal space is filled with fibrous tissue, fat, loose areolar tissue, blood vessels, lymphatic vessels, lymphnodes and nerves.

| Kidneys | Abdominal aorta |
|---|---|
| Adrenal glands | IVC |
| Ureter | Iliac vessels |
| Bladder | Spermatic and ovarian vessels |
| Ascending and descending colon | Renal vessels |
| Pancreas, Duodenum | Lumbar sympathetic chain |
| Seminal vesicles | Lymphatics and nodes |
| Vas deferens | Fatty areolar tissue |
| Upper rectum. & Vagina | |

## *LYMPHATIC DRAINAGE OF RETROPERITONEUM*



## *PREAORTIC NODES*

Lie directly anterior to the abdominal aorta.

They receive afferents from intermediate nodes associated with subdiaphamatic part of GIT, Liver, pancreas and spleen.

Their efferents form the intestinal trunk which enter the cisterna chyli.

## *LATERAL AORTIC NODES*

They lie on either side of abdominal aorta.

They receive afferents from the structures supplied by the lateral and dorsal branches of the aorta (Suprarenals, kidneys, testes and ovaries) and from the common iliac nodes.

Their efferents form a lumbar trunk on each side which terminate in Cisterna chili. Few efferents may pass to pre aortic nodes.

## *RETRO AORTIC NODES*

No particular drainage area

May be regarded as out lying member of the lateral aortic group.

# RETROPERITONEAL TUMOURS

## DEFINITION

Retropertitoneal tumours are tumours arising from the fat, muscle tissue, fibrous tissue, blood vessels, lymphnodes, nerves, and developmental remnants excluding tumours arising from retro peritoneal organs namely kidney, ureter, adrenal and pancreas.

## *CLASSIFICATION*

Retroperitoneal tumours can be classified as

1) Primary retroperitoneal tumours

2) Metastatic tumours

The most frequent tumours diagnosed in the retroperitoneum are :

- Retroperitoneal sarcomas
- Metastatic tumours
- Lymphomas
- Germ cell tumours
- Other neoplasms

CLASSIFICATION OF TUMOURS OF THE RETROPERITONEUM

| Tissue | Benign | Malignant |
|---|---|---|
| **Tumours arising from mesoderm :** | | |
| Adipose tissue | Lipoma | Liposarcoma |
| Fibrous tissue | Fibroma | Fiborsarcoma |
| Smooth muscle | Leiomyoma | Leiomyosarcoma |
| Striated Muscle | Rhabdomyoma | Rhabdomyosarcoma |
| Lymphatics | Lymphangioma | Lymphangiosarcoma |
| Primitive mesenchyme | Myxoma<br>mesenchymoma | Myxosarcoma |
| Histiocytes | Xanthogranuloma | Xanthosarcoma<br>Malignant fibrous histiocytoma |
| Blood Vessels | Hemangiopericytoma<br>Hemangioendothelioma | Malignant hemangio-pericytoma<br>Angiosarcoma |
| **Turmous arising from neural tissue** | | |
| Nerve sheath | Neurofibroma<br>Neurilemmoma | Neurogenic sarcoma<br>Malignant peripheral Nerve sheath tumour |
| Sympathetic nervous system | Ganglioneuroma | Neruoblastoma |
| Paraganglion system | Paraganglioma | Pheochromocytoma |
| **Tumours arising from embryologic remnants and heterotopic tissue:** | | |
| Embryologic remnants | Teratoma | Seminoma<br>Embryonal carcinoma<br>Teratoma, Endodermal sinus Tumour, Chordoma |
| Heterotopic adrenal tissue | | Wilm's tumours<br>Adrenocortical carcinoma |
| **Tumour arising from lymph nodes** | | Lymphomas<br>Secondary metastatic deposits |

**ETIOLOGY OF RETROPERITONEAL TUMOURS**

- Majority unknown
- Therapeutic radiation exposure
- Exposure to vinyl chloride, thorium dioxide and other agents.
- Associated with familial disorders like
    - Gardner's syndrome
    - Familial retinoblastoma
    - Neuro fibromastoses
    - Li-fraumani syndrome.
- Germ Line mutation of P53 gene.

## *PRIMARY RETROPERITONEAL TUMOURS*

- 75% of primary retroperitoneal tumours arises from mesoderm.
- 24% from ectoderm
- 1% from embryological remnants.
- Most retro peritoneal tumours are of mesodermal origin.
- Both benign and malignant tumours can arise from many different tissues.
- <25% tumours are benign. Most common benign tumours are lipoma and epithelial cysts.
- Retroperitoneal tumours can be cystic or solid in nature.
- Most of the benign tumours are cystic in nature.

- Most of the malignant tumours are solid in nature.
- Primary retro peritoneal tumours are mainly sarcomas, lymphomas and benign lesions.
- Retroperitoneal sarcomas represents 0.1-0.2% of all malignancies overall, only 10-15% of all soft-tissue sarcomas, approximately 40% of all retro peritoneal masses.

**1) TUMOURS OF ADIPOSE TISSUE**

Most frequently encountered mesenchymal tumours of retroperitoneum.

80% are abdominal with the perirenal area most frequently involved.

20% are pelvic.

**a) LIPOSARCOMA :**

More common primary malignant retro peritoneal tumour

More common than retro peritoneal lipomas

Adults commonly affected

Slightly increased incidence in females.

It is a tumour derived from undifferentiated mesenchymal cells, rather then the result of malignant transformation of normal retro peritoneal fat.

Histological varieties include
- Well differentiated
- Myxoid
- Pleomorphic
- Dedifferentiated

Well-differentiated and myxoid types have good prognosis. Pleomorphic and dedifferentiated types have poor prognosis.

Metastasis occur commonly to the lungs, liver and serosal surfaces (peritoneum),

infrequently to lymph nodes, mediastinum, heart.

Other tissue types may be found mixed with it leading to diagnosis of Firboliposarcoma,

Lipomyxosarcoma,

Fibromyxoliposarcoma.

Liposarcomas in other sites can occur simultaneously with, subsequent to or preceding retro peritoneal liposarcoma. They are considered multicentric tumours. Most have involved the lower extremities.

**b) LIPOMA**

- Less common than liposarcoma
- May contain fibrous, myxomatous, vascular elements leading to fibrolipoma, myxolipoma, fibromyxolipima.
- Adults predominantly affected
- Encapsulation is the rule, but may infiltrate or encompass adjacent structures.
- Recurrences following resection less common than in liposarcomas.

**c) HIBERNOMA**

- Uncommon, histologically distinct, benign adipose tumours
- Occurs in periadrenal, perirenal, para aortic area which are the sites where brown fat is found.

**II) TUMOURS ARISING FROM MUSCLE TISSUE :**

More frequent in women than men.

More frequent in adults than children

Types : Spindle cell variant

Epitheloid variant.

**a) LEIOMYOSARCOMA**

- Malignant tumour of smooth muscle
- More common than leiomyoma
- Origin of these tumours include several tissues located in the retroperitoneum, including blood vessels, spermatic cord, embryonic wolffian and mullerian duct remnants.

**b) LEIOMYOMA**

- Benign tumour of smooth muscle
- Exceptionally rare
- If encountered, consider uterine leiomyoma extending posteriorly, well differentiated leiomyosarcoma, lymphangiomyoma, angiomyolipoma.

**c) ANGIOMYOLIPOMA**

- Not strictly a primary retroperitoneal tumour
- Usually originates in the kidney. Rarely occurs in extra renal sites.
- Consists of mature fat & thick-walled blood vessel.
- Confused with leiomyosarcoma because of atypia seen in smooth muscle elements.

**d) RHABDOMYOSARCOMA**

- Malignant tumour of striated muscle
- Less common than leiomyosarcoma
- Embryonal, alveolar, mixed types occur in children
- Pleomorphic type in adults.

## *COSTELLO SYNDROME*

Children with mental retardation and retroperitoneal embryonal rhabdomyosarcoma.

**e) RHABDOMYOMA**

Rhabdomyoma in the retroperitoneum is very rare.

## *III. TUMOURS ARISING FROM FIBROUS TISSUE*

**a) BENIGN FIRBOUS TUMOURS**

- Previously called as fibromas. Now classified as fibromatosis.
- Generally solitary in one site; rarely, synchronous in the mesentery, retroperitoneum and abdominal wall scar.
- May be associated with familial polyposis, Gardner's syndrome, previous abdominal surgery.
- Distinguished from idiopathic fibrosis by lack of inflammatory cell component.
- Distinguished from fibrosarcoma by lack of cell anaplasia and mitosis.

**b) MALIGNANT FIRBOSARCOMA**

- Rarely reported
- May be associated with hypoglycemia

**c) INFLAMMATORY MYOFIBROBLASTIC TUMOUR**

Synonyms : Inflammatory pseudo tumour

Inflammatory fibro sarcoma.

It is pseudo sarcomtous proliferation, partially inflammatory in nature that occurs in soft tissues and viscera of children and young adults.

It can occur in the retroperitoneum.

It is now classified as intermediate fibrous tumour.

- Histopathologically composed of myofibroblasts and inflammatory cells.

**IV) TUMOURS ARISING FROM THE HISTIOCYTES :**

**a) MALIGNANT FIBROUS HISTIOCYTOMA**

- Are a group of sarcoma characterized histologically by storiform or cart-wheel like growth pattern of cells derived from tissue histiocytes. More common in adults than children, More common in males than females, more frequently in whites than blacks.
- Five Subtypes
    - Storiform pleomorphic
    - Myxoid
    - Gaint cell
    - Inflammtory
    - Angiomatoid
- Local recurrence and metastasis occur frequently to lungs.

V. TUMOURS ARISING FROM BLOOD VESSELS AND LYMPHATICS

a) BENIGN VASCULAR TUMOURS :

More common than malignant vascular tumours.

***LYMPHANGIOMA***

- Most common benign vascular tumours.
- Lymphangiectasia is the dilatation of abnormal lymphatic channels, which have failed to establish normal communication with the rest of the lymphatic system. The dilated lymphatic channels conglomerate and form a unilocular or multilocular cystic mass, known as lymphangioma.

- May occur as a part of syndrome consisting of chylous ascities and similar tumours in the lung or pleura or as a part of tuberous sclerosis.

OTHER BENIGN VASCULAR TUMOURS ARE:

Haemangiomas

Haemangioendotheliomas

Infantile haemangioendotheliomas, thrombocytopenia and bleeding is known as Kasabach Meritt Syndrome.

Benign hemangiopericytomas :

Derived from the pericytes. Present usually as bulky silent tumours.

b) MALIGNANT VASCULAR TUMOURS :

- less common than benign vascular tumours
- They include
    - Malignant hemangiopericytoma
    - Angiosarcoma

May be associated with hypoglycemia due to production of IGF by the tumours. IGF and IGF-R mRNA can be identified in tumour cells even in the absence of clinical hypoglycemia. Symptoms ablate with tumour removal.

## VI. OTHER MESENCHYMAL TISSUE TUMOURS :

### a) MYXOMAS :

**Contain abundant myxoid stroma and stellate cells mimicking primitive mesenchyme.**

Differentiated from myxoid liposaroma by lack of florid delicate vascularity.

b) MESENCHYMOMA :

Reported only in the retropeirtoneum and mesentry.

c) MYXOSARCOMA :

Malignant counterpart of myxoma.

**VII. TUMOURS ARISING FROM NERVE TISSUE :**

a) NEUROBLASTOMA :

- Most common malignant neurogenic tumours.
- Occurs exclusively in children.
- Extra-adrenal tumours half as frequent as adrenal origin.
- Generally solitary. May be multiple.
- Prognosis is better in young children and extra adrenal tumours.

b) GANGLIONEUROMA :

Affects old age group.

Extra-adrenal tumours ate more common than adrenal sites.

c) MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR :

- It is a malignant tumour arising from a peripheral nerve sheath.
- Histological types :

  Spindle cell

  Epithelioid

  Combined

- Unless the tumour is found arising from a nerve, it may be misinterpreted as fibrosarcoma or leiomyosarcoma.

d) NEUROFIBROMA :

e) NEURILEMMOMA :

Occurs in the retroperitoneum rarely.

Recurrence is common following resection.

f) PARAGANGLIOMA :

- Tumours of embryological origin arising from the neural crest.
- They can be found in any location along the aorta commonly in the region of organ of zukerkandal or in association with the sympathetic chain.
- These tumours can be non-functioning or functioning.
- Only 20% of paragangliomas are catecholamine secreting and cause a syndrome similar to that of pheochromocytoma.
- They may be multiple and malignant.
- Malignant type spread by blood stream predominantly to bones and lungs.

Some are familial (hereditary paraganglioma) and occur as a part of VHL Von Hippel Lindau disease.

CARNEY'S TRIAD :

- Multiple gastric stromal tumours,
- Pulmonary chordroma,
- Parganglioma.

g) CARCINOID TUMOUR :

It can arise in the retroperitoneum.

But, it may be metastasis from an undetected primary,

expression of a mesodernal teratoma,

neoplasm from endodermal cells normally present in this location.

VIII. RETROPERITONEAL EXTRAGONADAL GERM CELL TUMOURS :

- Primary tumours of extragonadal origin are rare.
- Retropertitoneum is the second most common site of extragonadal germ cell tumours.
- Two school of thoughts exist as to the origin of these neoplasms.
  - i) Displacement of primitive germ cells during early embryonic migration from the yolk sac ectoderm.
  - ii) Persistence of pluripotent cells in sequestered primitive rests during early somatic development.
- Distinction between primary EGCT and metastasis from an undetected primary testicular tumours may be difficult which has to be carried out by careful physical examination, supplemented by the use of high-resolution ultra sonogram.
- Both seminomas and non-seminomatous germ cell tumours can occur in the retroperitoneum.

Non-seminomatous GCT include embryonal carcinoma.

Yolk sac tumours, endodernal sinus tumour, teratoma.

Seminomas GCT are common in elderly males.

Non-seminomatous GCT occurs predominantly in female children.

- These tumours lack encapsulation, in contrast to their testicular counterparts, and tend to invade or envelope contiguous structures.

**IX. RETROPERIOTONEAL LYMPHOMA :**

- Retroperitoneum is a common location for lymphoma. Both Non-Hodgkin's lymphoma and Hodgkin's lymphoma can occur in the retroperitoneum.
- In children 30% have primary abdominal presentation. In adults this type of presentation is less common and generally it is part of a generalized involvement rather than being the only site of involvement.
- Retroperitoneal locations include para aortic, para caval, interaorto caval, renal hilar, supra hilar regions.
- Lymph nodes are said to be abnormal

  i) When enlarged in size > 1 cm in diameter in short axis.

  ii) When increased in number.

  iii) When characterized by aberrant internal architecture.

  Lymphomatous nodes will elevate the aorta. They can obstruct the infrior vena cava and can invade the pelvicalyceal system causing obstructive uropathy.

X. TUMO0URS ARISING FROM HETEROTOPIC TISSUE :

1. Extrarenal Wilm's tumour.
2. Heterotopic adrenocortical carcinoma.

Adrenal or renal cell type undifferentiated adenocarcinoma not involving adrenal gland or kidney can arise from heterotopic adrenal tissue and metanephric blastema.

XI. TUMORS RARELY ENCOUNTERED :

- Extraskeletal Ewing's Sarcoma.
- Mesothelioma
- Chondrosarcoma
- Clelar Cell Sarcoma
- Synovial Sarcoma

XII. TUMORS OF UNCERTAIN HISTOGENESIS :

- Granular cell myoblastoma
- Alveolar soft part sarcoma

XIII. RETROPERITONEAL CYSTS :

- majority of retroperitoneal cysts are benign in nature. They include,
- Cystic lesion arising from developmental remnants of the urogenital tract.
- Mesenteric cysts
- Teratomatous cysts
- Lymphogeneous cysts
- Serous & mucinous cystadenomas
- Malignant retro peritoneal cystic lesions include serous & mucinous cystadenocarcinomas.

MUCINOUS CYSTADENOCARCINOMAS :

The immunological staining characteristics of these malignant lesions have patterns similar to ovarian mucinous tumors. This genotypic similarity may indicate similar mechanisms in their histogenesis.

CLINICAL FEATURES OF REROPERITONEAL TUMOUS :

- Signs and symptoms of retroperitoneal tumors are vague.
- Present only when the tumour has attained a large size or by pressure on or infiltration of adjacent structures. This is because retroperitoneum is a potential space and most retro peritoneal tumours are expansile growths rather than aggressive infiltrating growths that produce early clinical problems and lead to investigation early in the course of the disease.
- Majority have vague abdominal pain/back pain, with a palpable abdominal/pelvic mass.
- Constitutional symptoms such as anorexia, weight loss, fever, fatigue, vomiting may be present.
- Advanced cases may present with

  * Features of caval compression –

    lower limb edema,

    varicocele,

    ascites,

    dilated abdominal wall veins.

  - Genitourinary symptoms
  - Venous thrombosis
  - Pleural effusion
  - Peripheral nerve disorders
  - Intra-abdominal bleeding
  - Highly vascular tumours may sequester platelets ands produce bleeding disorders.

Patients may present with features due to paraneoplastic syndrome like

Liposarcoma → Intermittent hypoglycemia

Germ cell tumour → Pre-cocious puberty

Neurobastoma → Opsoclonic myoclonus

Extra Adrenal Paraganglioma → Symptoms of Catecholamine excess.

CLINICAL EXAMINATION.

- Non tender, firm abdominal mass
- Liver
- Regional lymphnodes
- Scrotal examination

## *SPREAD OF RETROPERITONEAL TUMOURS*

Local Spread : Tumour invading along multiple tissue planes, blood vessels, nerves.

Hematogeneous Spread : Organs involved in the decreasing order of frequency are lung, liver, bone, brain.

Lymphatic Spread : Rare < 5% Embyonal rabdomyosarcoma

Lymphangiosarcoma

Epithelial sarcoma

## *IVESTIGATIONS*

**Lab Investigations** :

- Blood Urea, creatinine. Useful to assess renal function.
- LFT
- Turmour markers AFP, B-hcG. To R/o germ cell tumours.
- Complete hemogram, peripheral smear useful in cases of lymphomas.
- Plain X-Ray : Shows soft tissue shadow, shifting of bowel loops, calcification if any.

INTRAVENOUS UROGRAM (IVU)

To assess the anatomical and functional status of kidneys as removal of one kidney is often required for curative resections, any pressure effect over ureter and its effects.

Contrast CT obviates the need for IVU.

## *BARIUM MEAL SERIES AND BARIUM ENEMA*

Demonstrates displacement or invasion of bowel. May be occasionally needed in case of non-availability of CT.

ULTRASONOGRAM (USG):

Provides inadequate information in 30% due to interference from intraluminal gas. It may show mass, solid or cystic and secondary changes in the urinary tract.

Contrast enhanced (oral & intravenous) CT abdomen (CECT) :

- Best imaging modality for assessing retroperitoneal lymphadenopathy.
- Shows exact site and size of the tumour.
- Depicts anatomic changes secondary to its growth
- Tumour invasion of adjacent structures demonstrated or suggested

- Most retroperitoneal tumours appear as soft tissue masses with focal areas of necrosis but does not predict the histologic type or grade of sarcoma.
- Contrast CT obviates IVP
- CT guided core biopsy, FNA can be taken.

## *MRI MAGNETIC RESONANCE IMAGING*

Its advantages over CT scan are,

- Spatial assessment is better due to multiplanar capability.
- Clear view of vascular structures obtained without contrast.
- Demonstrates the extent of recurrent tumour
- Identifies the presence (or) absence of lymphadenopathy.
- Greater accuracy of defining tumour extent and respectability.

POSITRON EMISSION TOMOGRAPHY (PET SCAN)

- It is complementary to conventional staging modalities for staging.
- It is helpful in differentiating benign from malignant soft tissue masses. A tumour – background ratio greater than 3 is highly predictive of malignancy.
- The degree of FDG uptake correlates with the grade of the sarcoma and can guide the biopsy to be taken from the region with the highest grade.
- It is useful in monitoring the response to therapy and in the evaluation of residual masses after therapy.

## *ARTERIOGRAPHY*

Reveals the extent of the lesion, tumours blood supply, displacement of major vessels or pressure effects.

Considered in large lesions.

INFERIOR VENACAVOGRAM (IV Gram)

It may show vena caval obstruction.

RETROPERITONEOSCOPY:

Useful to diagnose retropritoneal masses and to take visually guided biopsies from the more representative area.

Used when CT / USG guided biopsy fails to establish a definite diagnosis.

In future, retroperitoneoscopy with visually guided biopsy will replace CT guided biopsy.

## *CHEST X-RAY OR CT THORAX*

To assess pulmonary metastases, pleural effusion, assessing mediastinal lymphadenopathy.

## *AJCC STAGING FOR SOFT TISSUE SARCOMAS*

### *G - Histologic Grade*

**Gx** - Grade cannot be assessed

**G1** - Well differentiated

**G2** - Moderately differentiated

**G3** - Poorly differentiated

**G4** - Undifferentiated

### *T Primary tumour Site*

**Tx** - Primary tumour size can not be assessed

**T0** - No evidence of primary tumour

**T1** - Tumour < 5 cm

**T1a** - Superficial tumour

**T1b** - Deep tumour

**T2** - Tumour 5cm > greater

**T2a** - Superficial tumour

**T2b** - Deep tumour

### *N Regional Nodes*

**Nx** - Regional nodes cannot be assessed

**N0** - No regional lymph node metastasis

**N1** - Regional lymphnode metastasis

**M, Distant Metastasis**

**Mx** - Presence of distant metastasis cannot be assessed

**Mo** - No distant metastasis

**M1** - Distant metastasis present

## *Staging*

| | | | | |
|---|---|---|---|---|
| Stage I | G1-2 | T1a, 1b, 2a, 2b | No | Mo |
| Stage II | G3-4 | T1a, 1b, 2a | No | Mo |
| Stage III | G3-4 | T2b | No | Mo |
| Stage IV | Any G | Any T | N1 | Mo |
| | Any G | Any T | Mo | M1 |

## *TREATMENT OF RETROPERITONEAL SARCOMAS*

The significant advances in multimodality therapy of extremity sarcomas have not been matched by similar progress in the management of retro peritoneal sarcomas.

Retroperitoneal sarcomas have a poor outcome. reasons being,

- Inability to diagnose these tumours at an early stage.
- Rarely cause significant symptoms until they achieve large size and even then symptoms are generally vague and nonspecific.
- Close relation to major blood vessels.
- Post op. RT difficult.

**PREPARATION**

1) Adequate blood should be arranged
2) Consent for removal of adjacent organs
3) Consent for faecal / urinary diversion.
4) Preoperative full bowel preparation is necessary as limited resection of the colon or rectum is commonly required.

## *APPROACHES*

i) Transabdominal approach

ii) Retroperitoneal or flank approach

iii) Thoraco abdominal approach

## *I) TRANSABDOMINAL APPROACH*

Advantages :

- Allows enbloc resection of involved organs.
- Early control of the vascular supply to the tumour

## *II) RETROPERITONEAL OR FLANK APPRAOCH*

Advantages :

- Post operative ileus is brief.
- Continued use of GIT to deliver nutritional support.
- Avoids handling of intestines and hence adhesions.
- Intraoperative heat loss and fluid loss is less.
- Post operative pneumonia and atelectasis is less.
- Abscess if it develops is interstitial and not intraperitoneal.

Disadvantages :

- Less satisfactory exposure in resection of adjacent organs.

### *III) thoraco abdominal approach*

It is indicated if the tumour is in the upper retroperitoneum or invading the diaphragm.

- Localised tumours should be removed enbloc with 1-2cm margins and enbloc resection of the involved organ most commonly the kidney, tail of pancreas or colon.
- Apparent capsule is usually a pseudocapsule containing normal as well as neoplastic cells. For curative resection, remove the tumour with surrounding clear margins. We should not remove the tumour from its pseudocapsule, because though dissection is easy, recurrence is almost certain.
- Retroperitoneal sarcomas are often fixed as they invade muscles of the posterior abdominal wall which are themselves immobile.
- Fixation is not a sign of unrestectability unless there is extensive involvement of irreparable or unremovable structures.
- Resectability rate varies between 30% - 100%.
- When retro peritoneal sarcoma is encountered unexpectedly at laparotomy, careful incisional biopsy with minimal disruption of surrounding tissue planes should be performed. Atrea of the biopsy should be isolated to prevent tumour spillage into the peritoneal cavity. When diagnosis is confirmed, wide excision carried out at the earliest.

**RESECTION RATE**

- It is 60% depending on the outlook of the surgeon.
- Highest resectability rate for liposarcoma and neurogenic sarcomas.

- Least resectability rate for leiomyosarcomas.
- For extensive low grade tumours, subtotal or palliative resections are beneficial.
- Complete resection rate 65% for Primary tumour, 44% for recurrent tumour.

## EXENT OF RESECTION

Resection of adjacent organs is necessary in 75% of patients for complete removal of retroperitoneal sarcomas.

Kidney and adrenals are the organs most commonly resected in curative resection followed by colon, pancreas, small bowel.

Radical lyphadenectomy is not indicated as the lymphnode metastasis is <5%.

**Partial Resection.**

- No survival benefit
- Used only for pain relief (or) bowel obstruction.

OPERATIVE MORTALITY :

It should be less than 5% with modern anaethesia and support.

## *OPERATIVE MORBIDITY*

Depends on the extent of resection and adjacent organs resected.

Common problems are small intestinal and colonic ileus,

- Perforation,
- Fistulas,
- Abdominal abscesses.
- Local Recurrence rate 40% to 85%.

PROGNOSIS & SURVIVAL :

Prognosis depends on two important factors :

a) Completeness of tumour resection.

b) Histological grade of tumour rather than histologic type.

Complete resection provides long term survival.

5 year survival rate after complete resection is 32-100%.

60-90% for low grade tumours,

15-50% for high grade tumours,

Overall survival including partial resection is 10%-50% at 5 years, 10%-20% at 10 years.

### *Based on histological grade*

If low grade - Recurrence occurs late, better survival

If high grade - Recurrence occurs early, poor survival

Retroperitoneal sarcomas are rarely totally cured because survival intervals, even if totally resected continue to decline beyond 15 years.

Recurrences occur in 50% of patients often in the original tumour bed and often resectable. So, careful long term follow up and aggressive surgical resection of recurrences when complete resection is possible. Tumours often becomes less differentiated and more aggressive with each recurrences, with short tumour free intervals.

Median time for recurrence – 19 mo for high grade and 44mo for low grade.

CAUSE OF DEATH is mainly due to local invasive effects of the tumorus.

## *RADIOTHERAPY*

- It is used as an adjuvant treatment following surgery as it is not possible in majority to obtain tumour clearance by surgical resection alone.
- Normal tissue tolerance to radiation is much lower in the abdomen and retroperitoneum than in the extremities. Extremity tumours are treated with 6000 cGY or more. In contrast, the small bowel can tolerate only 4500-5000 cGYand the liver and kidney, even less.
- In order to limit tissue toxicity, IORT (Intra Operative Radiotherapy) has been used. Here, the sensitive structures are moved out of the field while a single high dose or radiation is administered directly to the tumour bed. IORT is costly, logistically difficult and has its own complications such as neurotoxicity and available only in selected centers.

**RADIATION SENSITIZERS :**

- As the normal tissue tolerance of the retroperitoneum is limited, radiation sensitizers are being investigated to improve the effectiveness of EBRT (External Beam Radiotherapy).
- Iododeoxyuridine (IUDR) is the commonly used radiosensitizing agent. It is 100 times more effective than doxorubicin as a radiation sensitizer in vitro, although it does not have doxorubicin's direct cytotoxic effect.
- Studies of IUDR and EBRT in unresecetable sarcomas document impressively high rates of local control without surgery.
- Preoperative treatment with IUDR & radiation allows complete excision of gross tumour for most patients with retroperitoneal sarcomas and toxicity of this protocol has been acceptable.

CHEMOTHERAPY :

- Post operative adjuvant chemotherapy is of no benefit for retro peritoneal sarcomas and is poorly tolerated by patients, who have undergone a major intra-abdominal procedure with resection of multiple organs.
- Use of granulocyte macrophage colony stimulating factor has been associated with good response.
- Preoperative intra arterial chemotherapy is limited by the absence of a single feeding vessel in most retro peritoneal tumours.
- Complete Response rate of 15-35% have been reported with the use of adriamycin as a single agent.

TREATMENT OF RETROPERITONEAL GERM CELL TUMOURS

It is dependent on both the histologic type and size of the tumours.

Seminomas < 5cm, no metastasis → Radiotherapy 18-40 Gy.

Seminomas > 5cm → Initially chemotherapy, irradiate residual masses after chemotherapy.

Benign teratomas → Surgical resection.

Non-seminomastous Germ Cell tumours → Initially chemotherapy. Residual masses resected surgically.

**CHEMOTHERAPY REGIMENS :**

| | | | |
|---|---|---|---|
| PVB - | Cisplatin | VAB-6 - | Vinblastine |
| | Vinblastine | | Actinomycin –D |

| | | | | |
|---|---|---|---|---|
| | | Bleomycin | | Bleomycin |
| | | | | Cisplatin |
| BEP | - | Bleomycin | POMB - | Cisplatin |
| | | Etoposide | | Vincristine |
| | | Displatin | | Metotrexate |
| | | | | Bleomycin |
| | | | ACE - | Actinomycin D |
| | | | | Cyclophosphamide |
| | | | | Etoposide. |

## *METASTATIC RETROPERITONEAL TUMOURS*

Metastatic tumours in the retroperitoneum develops by two main routes.

i) Local extension.

ii) Lymphatic spread.

i) LOCAL SPREAD:

Local extension of tumour principally occurs in

Pancreatic carcinoma.

Primary bone tumours notably sacrococcygeal chordoma.

## *II) LYMPHATIC SPREAD*

Most common mode of spread to retroperitoneum.

Principal primary sites being.

Testes, Urinary bladder

Prostate Uterine cervix

Pancreas Endometrium

Kidney

Other primary sites include

Ovary Lung

Adrenals Breast

Stomach Melanoma

Colon

Diagnosis by USG, CT, FNAB.

Diagnosis based on presence of 'alien' calls, which are cells not normally indigenous to normal lymphnode constituents.

Cytologic appearance is similar to that described in their primary locations.

TREATMENT OF METASTATIC RETROPERITONEAL TUMOURS

Most of metastatic reroperitoneal tumours are treated by chemotherapy regimens depending on the primary site.

TREATMENT OF TESTICULAR METASTASIS TO RETROPERITONEAL NODES.

TREATMENT OF NON-SEMINOMATOUS GERM CELL TUMOUR METASTASIS

## *LOW STAGE DISEASE*

Stage I $T_{1-3}$ N0 M0

Stage IIA $T_{1-3}$N1M0

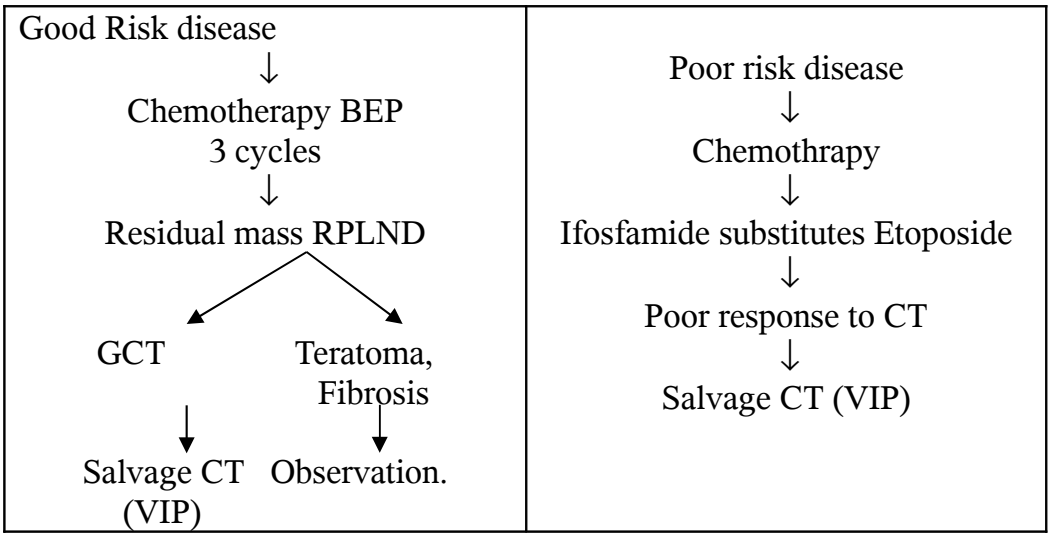
Stage II B $T_{1-3}$N2M0

- Retroperitoneal lymph node dissection (RPLND) followed by adjuvant chemotherapy BEP 2 cycles. (or) Primary Chemotherapy BEP for 3 cycles.

## *HIGH STAGE DISEASE*

Stage II C T1 N3 M0

Stage III Any T any N M1




**TREATMENT OF RETROPERITONEAL NODES FROM SEMINOMA**

Stage I : Abdominal radiotherapy 25 Gy to para Aortic nodes.

Stage II A, B : Adbominal radiotherapy 150 cGy per day 5 days per week for 6 weeks. Chemotherapy if involved nodes are close to kidney.

Stage IIIc, III : Chemotherapy (BEP) Post chemotherapy residual mass if > 3 cm surgical resection.

## *CHEMOTHERAPY REGIMENS*

BEP regimen

B Bleomycin - 30 U iv day 2,9,16

E Etoposide - 100mg/m2 iv day – 5

P cisplatin - 20mg/m2 iv day 1-5

Ifosfamide - 1.2g/m2 iv day 1-5

RETROPERITONEAL LYMPHNODE DISSECTION (RPLND)

- This is a highly sensitive though rigorous method of detecting involvement of retro peritoneal lymphnodes.
- Predominantly used in non-seminomaous tumors, because 20-30% of testicular non-seminomas with normal CT abdomen have RPLN metastases.
- Risk of metastases can be predicted by the vascular invasion in the primary tumour.
- The extent of lymph node involvement is predictive for relapse at other sites and can be used as a basis for considering adjuvant chemotherapy.

## *TECHNIQUES*

1) Bilateral RPLND : Suprahilar, infrahilar
2) Modified RPLND : For right and left testicular tumours.
3) Nerve sparing RPLND.

Surgical principles critical to perform RPLND safely :

i) Indepth understanding of retro peritoneal anatomy and be able to recognize common variations and their implications.

ii) Excellent exposure of the retroperitoneum.

iii) Thorough lymphadenectomy using ‘split & roll’ technique.

**APPROACHES**

| | | |
|---|---|---|
| 1) | Transabdominal | This approach is most commonly employed |
| 2) | Thoracoabdominal | This approach is used for suprahilar dissections. |
| 3) | Laparascopic | This approach is the noval developing technique. |

**Laparoscopic RPLND :**

This technique is used for Clinical Stage I and II testicular cancers. And it is found to be superior to open surgery in terms of surgical efficiency, morbidity and costs, whereas the therapeutic efficiency is equal.

## *SPLIT AND ROLL TECHNIQUE*

Allows enbloc removal of the nodes. The lumbar vessels must be divided twice, first at the wall of the great vessels and again as they enter the foramina alongside the vertebral bodies.

Important aspect of performing nerve-sparing RPLND

Identification and preservation of

i) Sympathetic chains bilaterally

ii) Postganglionic sympathetic nerves arising from the sympathetic chains.

iii) Hypogastric plexus which is the anastomosing network of nerve fibers anterior to lower aorta.

The most important nerves are those arising from L3 and L4 ganglion, which are essentials for preserving antegrade ejaculation.

## *COMPLICATIONS OF RPLND*

Major complications include

- Small bowel obstruction
- Lymphocele
- Wound dehiscence

Pulmoary complications in thoracoabdominal approach which include atelectasis, prolonged chest tube drainage, need for increased post operative analgesia.

Loss of antegrade ejaculation and consequently potential infertility.

It is inevitable after the bilateral RPLND

It is reduced by nerve sparing RPLND.

## *RETROPERITONEAL LYMPHOCELE*

- It is a post operative complication following RPLND.
- Usually develops within 10-21 days of surgery
- It can occur in both retroperitoneum and peritoneal compartments but retroperitoneal location is more commion.

Investigations :

CT & MRI : Shows encapsulated fluid collections fo varying complexity.

Distinction between lymphoceles, cystic tumour recurrence and other abnormal fluid collections can be difficult.

USG : Anechoic appearance similar to simple cysts.

Distinction from an abscess, hematoma, urinoma, fluid – filled bowel is often difficult, especially if internal debris is present.

Because of poor specificity of CT, MRI, USG ; percutaneous aspiration is often required for definitive diagnosis.

Treatment :

Small anechoic lymphoceles often resolve spontaneously.

Large, echogenic lymphoceles may require

- Drainage
- Sclerosing therapy
- Surgical resection.

## *TREATMENT OF LYMPHOMAS*

Modified Ann Arbor Staging for Hodgkin's Lymphoma

**Stage I** : Single lymphnode region (1) or one extra lymphatic site (1E)

**Stage II** : Two (or) more lymphnode regions, same side of the diaphragm (11) or local extralymphatic extension plus one (or) more lymphnode regions on same side of diaphragm (11E).

**Stage III** : Lymphnode regions on both side of the diaphragm (111), which may be accompanied by local extra lymphatic extension (IIIE) (or) Spleen (111s).

**Stage IV** : Diffuse involvement of one (or) more extra lymphatic organ (or) site.

**B Symptoms** : Presence of at least one of the following.

1) Weight loss > 10% during 6 months
2) Recurrent unexplained fever > $38^{O}C$
3) Recurrent night sweats

## *OVERVIEW OF TREATMENT OF HODGKIN'S LYMPHOMA*

| **Stage Ia, IIa** | **Low Bulk** | **ABVD x 2 + IRRT** |
|---|---|---|
| Any stage with B symptoms (or) stage III or IV | Low Bulk | ABVD untill 2 cycles past complete response (Min – 6 cycles Max – 8 cycles.) |
| Any Stage | Bulky | ABVD x 6 +_ IRRT |

IRRT – involved Region Radiotherapy

Bulky - ≥ 10cm largest diameter of any single mass or mediastinal mass ratio > 1/3 largest transthoracic diameter.

**CHEMOTHERAPY REGIMNS USED FOR HODGKIN'S LYMPHOMA**

**1) ABVD**

A: Adriamycin – 25mg / sq.meter IV 1 & 15

B : Bleomycin – 10 units / sq. meter IV 1 & 15

V : Vinblastine – 6mg / sq. meter IV 1 & 15

D : Dacarbazine 375mg/ sq. meter IV 1 & 15

Repeat every 28 days for 6 cycles

**2) MOPP**

M : Mechlorethamine – 6mg / sq. meter IV 1 & 8

O : Oncovin – 1.4mg / sq. meter IV 1 & 8

P : Pro carbazine – 100mg / sq. meter PO 1-14

P : Prednisolone – 40mg / sq. meter PO 1-14.

Repeat every 28 days for 6 cycles.

**3) MVPP**

Mechlorethamine, Vinblastine, Procarbazine, Prednisolone

**4) ChlVPP**

Chlorambucil, Vinblastine, Procarbazine, Prednisolone.

## *TREATMNT OF NON-HODGKIN'S LYMPHOMA*

**CLASSIFICATION OF NHL**

### *I) Low Grade NHL*

- Small lymphocytic
- Follicular predominantly small cell type
- Follicular mixed cell type

### *II) Intermediate Grade NHL*

- Follicular predominantly large cell type
- Diffuse – small cell, large cell (or) mixed type

### *III) High Grade NHL*

- Large Cell immunoblastic
- Lymphoblastic
- Burkitt's lymphoma

## *TREATMENT OF LOW GRADE NHL*

**Stage I & II** – Radiotherapy

3600 – 4500 cGY

100 – 150 cGY / day for 5 days / wk for 6 wks

Limited abdominal NHL – Para aortic radiation.

**Stage III & IV** – Chemotherapy.

## *TREATMENT OF INTERMEDIATE AND HIGH GRADE WHL*

For all Stages – Chemotherapy

CHOP Regimn

C : Cyclophosphamide 750mg / sq. meter IV on day 1

H : Doxarubicin 50mg / sq. meter IV on day 1

O : Oncovin 1.4mg / sq. meter IV on day 1

P : Prednisolone 100mg orally for days 1-5

Repeat every 21 days for 6 cycles.

## *ROLE OF SURGERY IN RETROPERITONEAL LYMPHOMAS*

If lymphomas found accidentally on laprotomy

If localized resect fully.

If diffuse, biopsy alone taken and margins of tumour marked by clips.

# MATERIALS AND METHODS

The clinical material used in this study consists of 26 patients of suspected retroperitoneal tumours in Govt. Stanley Hospital during the period from July 2003 to March 2006.

## *MATERIALS*

1) Clinical evaluation of

Age and sex incidence

Presenting symptoms

Clinical examination.

2) Investigations

Routine haematological and biochemical investigations

Peripheral smear

HIV – ELISA test

Chest X-ray

USG abdomen

Contrast CT abdomen

IVU

Lymph node biopsy

Laparoscopy

BMFT / BA enema

Colonoscopy

## *METHODOLOGY*

All patients are thoroughly examined and necessary investigations done.

Treatment modality is then planned according to the investigation report and are managed accordingly.

Surgically resected specimens are sent for histopathological examination.

Patient immediate post operative period was studied.

# OBSERVATION AND RESULTS

The following observations were made in our study on retroperitoneal tumours.

DISTRIBUTION OF DIFFERENT TYPES OF RETROPERITONEAL TUMOURS

| Type | Number | Percentage |
|---|---|---|
| Primary Retro peritoneal tumours | 16 | 61.5 |
| Retroperitoneal lymphomas | 6 | 23.1 |
| Secondary metastasis | 4 | 15.4 |
| Total | 26 | |

Primary retro peritoneal tumours are the most common type followed by retro peritoneal lymphomas and secondaries.

## AGE INCIDENCE OF RETROPERITONEAL TUMOURS

| Age (yrs) | Number | Percentage |
|---|---|---|
| 20-30 | 6 | 23.1% |
| 30-40 | 8 | 30.8% |
| 40-50 | 4 | 15.4% |
| 50-60 | 6 | 23.1 |
| 60-70 | 2 | 7.6 |

54% of retro peritoneal tumours occurred in age group of 20-40 years.

## SEX INCIDENCE OF RETROPERITONEAL TUMOURS

| Sex | Number | Percentage |
|---|---|---|
| Male | 11 | 42.3% |
| Female | 15 | 57.7% |

The male : female ratio was 1:1.36.

CLINICAL PRESENTATION OF RETROPERITONEAL TUMOURS

| **Symptoms** | **Number** | **Percentage** |
|---|---|---|
| Incidental | 2 | 7.7 |
| Abdominal mass | 24 | 92.3 |
| Abdominal Pain | 15 | 57.7 |
| Loss of Weight | 22 | 84.6 |
| Loss of Appetite | 12 | 46.1 |
| Urinary Tract symptoms | 8 | 30.8 |
| GI Symptoms | 2 | 7.7 |
| Oedema / Pain | 2 | 7.7 |
| Emergency | - | - |

92% of patients presented with abdominal mass.

Other commonly noted symptoms are pain, loss of weight and loss of appetite.

## DISTRIBUTION OF RETROPERITONEAL LYMPHOMAS

| Type | As part of Gen. Lymphadenopthy | Localised retro peritoneal involvement |
|---|---|---|
| Hodgkin's | 1 | - |
| Non-Hodgkin's | 4 | 1 |
| | Total | 6 |

Majority of retro peritoneal lymphomas are of non-Hodgkin's type occurring as a part of generalized lymphadenopathy.

## INVESTIGATIONS DONE

| Investigations | Number |
|---|---|
| USG – Abdomen | 26 |
| Contract CT abdomen | 26 |
| X-ray chest | 26 |
| Lymphnode biopsy | 6 |
| IVU | 8 |
| Barium enema | 2 |
| Laproscopy | 1 |
| Colonoscopy | 1 |

## TREATMENT OFFERED

| | Type of surgery | Number |
|---|---|---|
| Operative | Complete Excision of Tumour only | 6 |
| | Complete Excision of Tumour + (R) Hemicolectomy | 1 |
| | Complete excision of tumour + vascular graft | Nil |
| | Incomplete excision (Debulking) | 6 |

| | | |
|---|---|---|
| Non – Operative ↓ Chemotherapy | Lymphomas | 6 |
| | Metastatic Tumours | 4 |
| | Tumours found inoperable by investigations | 3 |

## POST OPERATIVE COMPLICATIONS

| Post op complications | Number |
|---|---|
| Prolonged Paralytic ileus | 2 |
| Wound infection | 1 |
| Death | 1 |
| Total | 4 |

One patient died due to respiratory complications.

## CLEARANCE IN SURGICAL RESECTED TUMOURS

| | |
|---|---|
| Total No. of Patients | 7 |
| Positive Tumour margin | 1 |
| Samples from bed positive | 2 |

3 patients had histologically positive margins and one of them presented 2 years later with recurrence.

## HISTOLOGY DISTRIBUTION

| | **Type** | **Number** |
|---|---|---|
| Primary | Liposarcoma | 9 |
| | Fibrosarcoma | 4 |
| | Leiomyosarcoma | 1 |
| | Neuro ecto dermal tumour | 2 |
| Secondary | Granulosa cell tumour of ovary | 1 |
| | Adenocarcinoma | 3 |
| Lymphomas | Non-Hodgkin’s | 5 |
| | Hodgkin’s | 1 |

Majority of the retroperitoneal tumours are of primary type of which liposarcoma is the commonest followed by fibrosarcoma.

Of the secondaries, adenocarcinoma was the commonest type in our study.

SELECTED REPORTS EVALUATING SURGICAL TREATMENT OF PRIMARY RETROPERITONEAL TUMOURS

| Study | No. of Patients | Complete Resection | 5 – year Local Recurrence | 5 – year survival |
|---|---|---|---|---|
| Lewis et al (1998) | 500 | 80% | 59% | 70% |
| Jagues et al (1990) | 1143 | 65% | 49% | NR |
| Stoeckle et al (2001) | 165 | 65% | 48% | 46% |
| Hassan et al (2004) | 97 | 78% | 44% | 51% |
| Alvavenga et al (1991) | 120 | 25% | 80% | 29% |
| Sunger et al 1993 | 83 | NR | NR | 54 |
| Karakonsis et al (1996) | 57 | 100 | 42 | 66 |

**ALJEIRAN RA, LOPEZ GC, HERVA GA ET AL.**

Rev Inst. Nal. Cancercol. (Men) 1977 43(4) 194-199.

Most common histology : Liposarcoma 55%. Leumyosarcoma 16%.

Complete resection rate 71%

Partial resection rate 11%

**CHRISTOPHER WONDLHAM, Peter WT Pristel et al**

Cancer control Jan / Feb Vol. 12, No.1 (35/36)

Most common histology

Lipo sarcoma 41%

Lecomyosarcoma 28%

MFH 7%

Fibrosarcoma 6%

MPNST 3%

Most common presentations : Abdominal mass.

The incidence is equal between both sexes.

**BURTON L. ESENBERG ET AL**

State of the science June 2002.

Most common histology liposarcoma 40%

Leiomyosarcoma 30%.

Complete resection rate 50-60%.

**LEWIS JJ LAING D, W ORDEROFF JM, BRENNAN MF ET AL**

Dept. of Surgery

Memmorial Sloan Kettering Cancer Centre, New York.

Most common histology : MFH : 40%

Liposarcoma : 25%

**TORNI FERRARIO, CONSTANTINE P KARAKOUSIS ET AL**

Archives of Surgery Vol. 138, No.3, Mar. 2003.

Complete resection rate 95%

99% primary tumours

90% for recurrent tumours

Local recurrence 41%.

# OBSERVATION AND ANALYSIS

In this study of 26 patients with retroperitoneal tumours following observations were made.

1. Majority of the retroperitoneal tumours occurred in the age group 20-40 years.

2. The incidence was almost equal in both male and females. This is consistent with the study conducted by Christopher Wondlham, Peter WT prisstel et al. Cancer control Jan / Feb Vol. 12, No.1 (35/36)

3. Primary retro peritoneal tumours are the most common type followed by retro peritoneal lymphomas and then secondaries.

4. Abdominal mass was the most common initial presentation. This is consistent with the study conducted by Christopher Wondlham, Peter WT prisstel et al. Cancer control Jan / Feb Vol. 12, No.1 (35/36)

5. Majority of retroperitoneal lymphoma are of non-Hodgkin's type.

6. Surgery was the primary modality of treatment in retroperitoneal sarcomas and chemotherapy was given to retro peritoneal lymphomas and secondaries.

7. Liposarcoma was the most common histological subtype encountered in our study. This is consistent with the studies conducted by Aljeiran Ra, Lopez Gc, Herva Ga Et Al, Christopher Wondlham, Peter WT Pristel et al, Burton l. Esenberg et al.

8. The complete resection rate in our study was 53.8%. This is less than the studies conducted by Lewis et al, Jagues et al but similar to the studies conducted by Burton L. Esenberg et al.

**DISCUSSION**

Retroperitoneal tumours are relatively rare accounting for less than 1% of tumours encountered in our hospital.

## *RETROPERITONEAL TUMOURS PRIMARY*

Primary retro peritoneal tumours accounts for 61.5% of retroperitoneal tumours.

Majority of them presented with abdominal mass followed by loss of weight and loss of appetite and pain.

Two patients had symptoms of sub acute intestinal obstruction and another two had pain radiating down the leg.

USG abdomen and contrast CT abdomen was done in all patients.

Three patients had growth closely related to major vessels and were decided unresectable.

Seven Patients had complete excision of the tumour mass and debulking was done in 6 patients and were followed up with radiotherapy.

In one patient, resection and anastomosis of small bowel and Rt hemicolectomy was done along with the tumour.

Majority of the tumours were liposarcoma, the next common being fibrosarcoma.

One patient died on 17$^{th}$ post operative day die to respiratory complications.

## *RETROPERITONEAL LYMPHOMA*

Majority of the retroperitoneal lymphomas are of NHL, intermediate grade type.

Lymphnode biopsy was done in all cases to confirm the diagnosis.

Complete haemogram, peripheral smear, ELISA, USG abdomen and contrast CT abdomen were also done.

Five patient had non-Hodgkin's lymphoma and were treated with CHOP regimen chemotherapy.

One patient had Hodgkin's lymphoma and was treated with MOP regimen.

Chemotherapy was found to be successful and majority of patients responded well.

### *METASTATIC RETROPERITONEAL TUMOURS*

Majority of retroperitoneal metastasis in this study were adenocarcinoma and were treated with chemotherapy according to the primary histology.

Most of the patients did not come for followup so recurrence, mortality and morbidity could not be assessed.

# CONCLUSION

In this study of 26 patients with retroperitoneal tumours following conclusions were made

1) Majority of the retroperitoneal tumours occurred in the age group 20-40 years.

2) The incidence was almost equal in both male and females.

3) Primary retroperitoneal tumours are the most common type followed by retro peritoneal lymphomas and then secondaries.

4) Abdominal mass was the most common initial presentation.

5) Majority of retroperitoneal lymphoma are of non-Hodgkin's type.

6) Surgery was the primary modality of treatment in retroperitoneal sarcomas and chemotherapy was given to retroperitoneal lymphomas and secondaries.

7) Liposarcoma was the most common histological subtype encountered in our study.

# PROFORMA

Name : Age : Sex : IP No.

Occupation : D.O.A. :

Address : D.O.S. :

D.O.D.:

## CHIEF COMPLAINTS AND THEIR DURATION

Abdominal mass

Abdominal pain

Loss of weight

Loss of appetite

Nausea and vomiting

Constipation

Fever

Urinary tract symptoms

Lower limb edema / pain

## PHYSICAL EXAMINATION

Built

Nourishment

Anaemia

Lymphadenopathy

Pedal edema

Swelling elsewhere

Café lait spots

Abdominal examination

Character of the swelling Site :

Size :

Shape :

**Plane :**

## INVESTIGATIONS

Routine blood and urine examinations

Complete haemogram

Peripheral smear

ELISA

X-ray chest

FNAC

Lymph node biopsy

USG – abdomen

Barium enema / colonoscopy

Contrast CT abdomen

**TREATMENT GIVEN**

**IMMEDIATE POST OPERATIVE FOLLOWUP**

**HPE REPORT**

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